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Co-morbidity in a cystic fibrosis population attending a regional clinic

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Aims: We noted a significant number of cystic fibrosis (CF) patients attending a regional clinic had additional co-morbid conditions including neoplasms, thus adding to their complex management.
Methods: In October 2004 we reviewed all 115 patients attending CF clinics in Limerick Regional Hospital – 42 adults and 73 attending paediatric services. We looked at the prevalence of additional medical diagnoses, excluding any recognised complications of CF.
Results: We noted 16 cases (14%) with co-morbid conditions. Most notably, 3 had neoplasms, including one 19 year old male with metastatic, poorly differentiated, adenocarcinoma of the ileocaecal valve. The second neoplasm was a stage 2 testicular teratoma in a 24 year old male. The third was a mature ovarian teratoma in a 4 year old girl.
In addition, 4 cases had renal problems including ureterocele, IgA nephropathy, duplex kidney and pelviureteric junction obstruction.
4 patients were found to have neurological disorders including 3 female patients with epilepsy i.e. partial seizures, absence seizures and juvenile myoclonic epilepsy. The fourth case was a female with a left hemiplegia secondary to a right middle cerebral artery infarct. Other conditions included 1 each of developmental dysplasia of the hip, atrioseptal defect, phimosis, hepatitis B and attention deficit hyperactivity disorder.
Conclusion: Of 115 patients attending a regional CF clinic, a significant number have co-morbid conditions including 3 with neoplasms. This has an additional impact on their complex management as well as prognosis and life expectancy.

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Lupus erythematosus and Cystic Fibrosis. Case report and screening in the CF-population

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We report about the rare association of Cystic Fibrosis and lupus erythematosus in a 49 years old woman genotype F508del/G85E and present the results of a screening for antinuclear antibodies in 50 CF-patients.
In our patient CF diagnose was made in 1987 by three positive sweat tests (Cl^- >100 mmol/l). At this time she had a history of recurrent pneumonia and pulmonary colonisation with mucoid Pseudomonas as well as moderate pancreatic insufficiency.
Since 1998 she complained about diffuse pain of muscles and joints. Initially shoulders and knees were involved; later she also suffered from pain and swelling of the fingers. Exposition to sunlight provoked skin rushes; symptoms of erythema nodosum revealed on forearms and knees.
Additionally to these clinical signs the following serum antibodies were found: ANA 1:5120, ds-DNA 203 IU/ml, ss-DNA 226 IU/ml, Histon antibodies 32 IU/ml and positive circulating immune complexes. The diagnose of lupus erythematosus was made as more than 5 of 11 diagnostic criteria defined by the American College of Rheumatology were fulfilled.
We screened our CF-population to evaluate, whether clinical or serological signs for lupus erythematosus occur frequently in Cystic Fibrosis.
Results: In a 33 years old patient high antinuclear antibodies were found (ANA 1:5120); two 17 and 38 years old patients had positive titres for ds-DNA (40.2 IU/ml/27.9 IU/ml); one female adult patient suffered from CF arthritis. All of these patients did not reveal further clinical or serological criteria of lupus erythematosus.
Conclusion: We present the rare association of CF and lupus erythematosus not described in literature. The results of our screening point out the need of further investigation in this field.

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Cystic fibrosis and cancer relation

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The permanent improvement of the management of CF, leads to a significant increase in the quality and life expectation in these patients. In association to the well-known clinical polymorphism of the patients with CF a new problem is raising, respectively higher incidence of cancer, in general, especially of some particular forms. The aim of study was the evaluation of the risk cancer in CF patients from Romania. Study method: retrospective study of 20 CF adult patients (6 male and 14 female) from CF Centre Timisora. Results: At the time of study 5 cases deceased (mean age 21, 4 years) and 15 alive (mean age 23,1 years). The genotype of deceased cases was: homozygote DF508 at 3 patients and compound heterozygote genotype at 2 cases. The genotype of alive cases was: 3 cases with homozygote DF508, 10 cases compound heterozygote DF508 and 2 cases with DF508 and polymorphic allele. Mean age of our patients for the onset of a malignant status is superposed with literature data. Anyway specific investigations aiming for the detection of a form of cancer were negative in the studied lot. Conclusions: our results do not confirm the literature data, but we are not in the position to make any statistical considerations; remains for discussions the role of the exogenous factors, without a precise evaluation of these.

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A study to investigate the reliability of results of Tobramycin levels taken from a "port" in patients with cystic fibrosis

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Aims: To compare the results of Tobramycin blood levels from "ports" (totally implanted venous access devices) with those from capillary samples. If shown to be comparable, the use of "port" levels would save our patients another painful procedure. Levels of other drugs e.g. Vancomycin are determined using blood from "ports".
Methods: All patients with CF who were receiving intravenous Tobramycin three times daily via a "port" were eligible for the study. After informed consent was obtained, paired "port" and capillary/venous samples were taken at the appropriate time during the antibiotic course. Blood was taken from the catheter in situ immediately after the capillary sample was taken. Clinical decisions were taken on the capillary result as per normal. To avoid bias the "port" results were kept from the investigators making clinical decisions.
Results: The study was discontinued after 11-paired samples from 11 children were obtained. The results showed very different levels in the paired samples with no correlation within the pairs. It was deemed unethical to continue.
Conclusion: We conclude that using blood from a "port" is not a reliable method of checking Tobramycin levels.